

Sarcoidosis

Birendra Sah, MD, FCCP

Director, Sarcoidosis program

Assistant Professor of Medicine

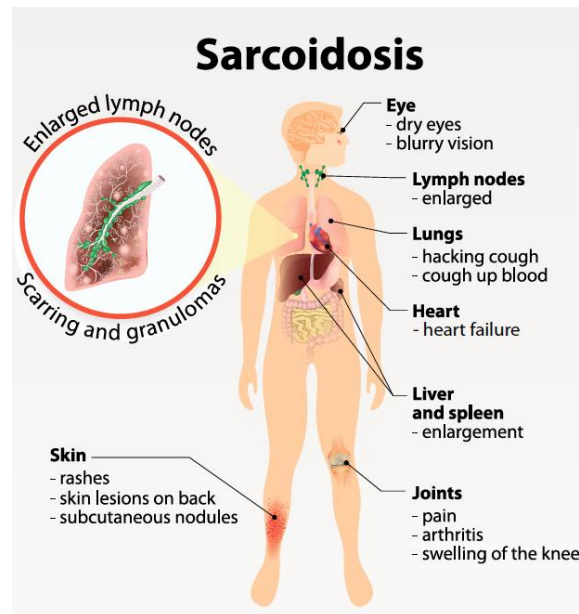
Pulmonary & Critical Care Division

Disclosure

- No financial conflict of interest

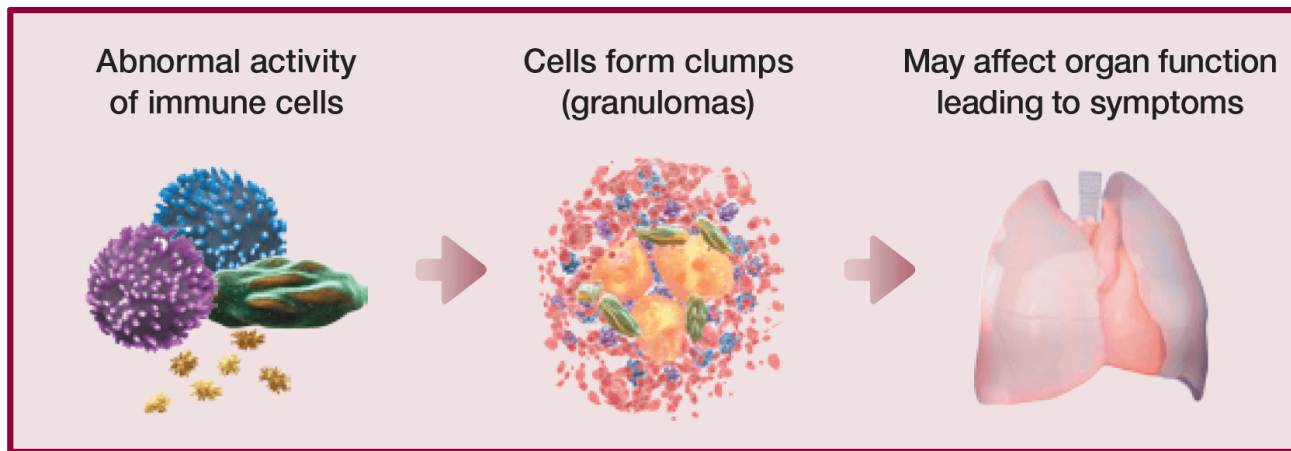
What is Sarcoidosis ?

- A chronic inflammatory disease which can affect any organ of body but most commonly lungs.



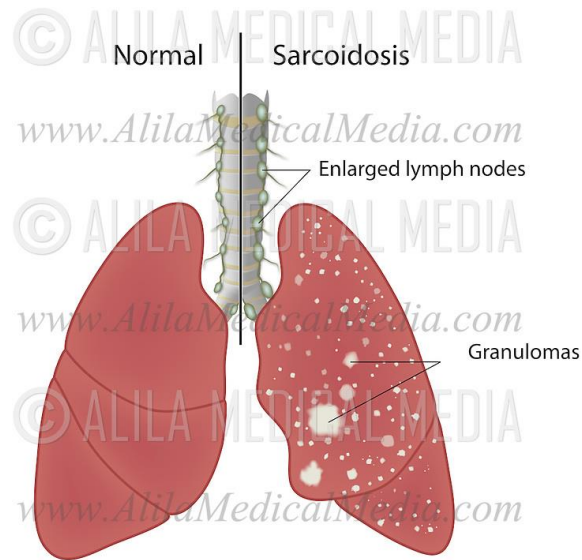
What is Sarcoidosis?

- Characterized by overreaction of immune system which results in the build up of several small clusters of inflammatory cells called “Granulomas” in the affected organs.



What is Sarcoidosis?

- Formation of too many of these clusters may interfere with organ structure and function.



© Alila Medical Media - www.AlilaMedicalMedia.com

Possible causes

- No one knows exactly what causes sarcoidosis
- Thought to be resulting from one or more exposures to “trigger factors” in people with certain genetic make up.

Possible causes

Suspected triggers

- bacteria,
 - viruses,
 - fungi
 - chemicals
- These triggers are usually harmless in most people, but may irritate the immune systems of people who are at genetic risk for developing sarcoidosis.

Risk Factors

- Affects people worldwide, of any age, race, gender
- In US, most common and severe in African Americans and European “particularly Scandinavian” descent.
- Age: 20 - 40 yrs of age
- Gender: Women > men

Associated Risk Factors

- Environmental exposure
 - inorganic particles, insecticides, and mold
- Occupational exposure
 - U.S. Navy, metalworking, firefighting, and the handling of building supplies
- Fire fighters and rescue workers involved in 2001 WTC disaster

Genetic Factors

- Risk higher if close family members have sarcoidosis

Symptoms

- Many people are often asymptomatic , detected incidentally on chest X-ray done during routine physical exam
- Symptomatic people can have variable signs and symptoms depending on the organs affected.

Symptoms

Common symptoms:

- shortness of breath,
- wheezing,
- chronic cough
- Chest pain

Other Symptoms

- Fatigue
- Weight loss
- Night sweats
- Skin rashes
- Eye symptoms
- Irregular heart beat
- Leg swelling
- Joint pain, swelling
- Muscle soreness
- Weakness & Numbness
- Enlarged Lymph nodes
- High blood or urine calcium level
- Kidney stones, Renal failure

Diagnosis

- Sarcoidosis is a diagnosis of exclusion
- There is no objective test which can easily diagnose sarcoidosis.
- Numerous exams and tests are required to confirm the diagnosis and to decide the best treatment options.

Diagnosis

- Chest X-ray
- CT scan
- Lung function test
- Biopsy of the affected organs (Lungs, lymph nodes, skin, bone, liver)
- Blood tests (Blood cell count, liver test, renal function test, blood and urine calcium, ACE level)
- MRI
- PET scan
- Cardiac tests (EKG, Echo, heart rhythm monitoring)

Clinical course

- In many cases of sarcoidosis, no treatment is necessary and sarcoidosis may go away in 2-5 years without medical treatment.
- The disease may never reappear or may reappear (in ~ 5%) later in life.
- In other patients, the disease is progressive, causing scarring in affected organs and requiring ongoing treatment

Treatment

- Treatment is not indicated in asymptomatic patients
- These patients are monitored closely for worsening
- Treatment is started when symptoms are severe enough to impair quality of life or when there are danger organ damage or life.

Medications

- **Corticosteroids: Prednisone**

- First line treatment
- Reduces inflammation
- Starting dose 20-40 mg daily
- Relieves symptoms in 1-3 months
- Then taper prednisone slowly to the lowest dose which will keep symptoms under control and cause less side effects
- Stay on this dose for another 6 months then taper it off
- Common Side effects: weight gain, insomnia, mood swings, acne, high blood sugar, osteoporosis, risk of infections

Medications

- **Immune system suppressant medication:**
 - **Methotrexate, Azathioprine, Leflunomide, Cellcept**
 - 2nd line treatment
 - Indicated when prednisone is not effective or causing significant side effects
 - Can take up to 6 months to show its effects
 - Side effects: Liver, kidney, bone marrow toxicities, malignancies
 - Blood tests are done every 1-3 months to monitor side effects

Medications

- **Antimalarial drugs:**
 - **Hydroxychloroquine (Plaquinil)**
 - Effective in skin symptoms or a high level of calcium
 - Side effects: Nausea, vision problem
 - Needs eye exam every 6 months

Medications

- **Biological agent (Anti-TNF)**
 - Infliximab (Remicade) IV
 - Adalimumab (Humira) SQ
 - Third line treatment
 - Side effects: Risk of infections, liver toxicities, lupus like illness, cancers

Treatment

- Sarcoidosis is often treated with the help of a multidisciplinary team of health care professionals who specialize in the treatment of the lungs, heart, brain, kidneys, liver, eyes, and skin.
- At specialized medical centers, these health care providers work as a team to develop a comprehensive treatment plan.

Follow up

- Asymptomatic patients
 - Initially every 6 months, then annually for 3-5 years
- Symptomatic patients
 - Every 3-6 months
- Follow up test:
 - Blood tests and PFT every 3-6 months
 - Chest X-ray every 6-12 months

Complications

- Fibrosis of lungs
- Cavities in lungs with fungal ball formation
- Pulmonary hypertension
- Side effects of medications
- Fatigue, depression, small fiber neuropathy

Take home points

- A chronic inflammatory disease characterized by formation of clusters inflammatory cells “granuloma”
- Exact cause unknown but is thought to be due to overreaction of immune system in response to exposure to triggers in people with certain genetic make up
- Has been associated with exposure to inorganic particles, insecticides, molds, certain occupations like navy, firefighters, metalworking,

Take home points

- More common and severe in AA
- Risk increases if close relatives have sarcoidosis
- Can affect any organ but most common in lungs
- Biopsy of affected organ is needed for diagnosis
- Many patients are asymptomatic and may go away in 2-5 years without medical treatment
- Common symptoms are respiratory followed skin, eye
- Treatment is indicated only in symptomatic patients or if affecting, heart, brain
- First line treatment : Prednisone

Thank you

- Questions?

Question

What is the definition of Sarcoidosis ?

